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Quality of life and sexual well-being in patients with a Fontan circulation: An explorative pilot study with a mixed method design

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Abstract

Objective: To get an impression of the quality of life (QOL) and sexual well-being in the Fontan population, and to generate hypotheses for future research.

Methods: For this cross-sectional pilot study, questionnaires regarding health-related QOL, sexual function and fertility/pregnancy were completed by 21 patients with a Fontan circulation >16 years old, followed at the University Medical Center Groningen, the Netherlands. Semi-structured qualitative interviews were conducted in 8 patients.

Results: Fontan patients scored significantly lower on general health than their healthy peers ($t(19) = -3.0$, $P = .008$), whereas their scores on other QOL domains and sexual well-being were comparable to normal values. During childhood, most patients experienced physical limitations and the feeling of being an outsider, and frequently faced bullying. Regarding sexual well-being, large inter-individual differences were noted. Four interviewed patients (25–30 years) reported a good sexual well-being, whereas the other interviewed patients (33–47 years) reported erectile dysfunction, low self-esteem and avoidance of sexual intercourse. Both the QOL domains mental health and role restrictions due to emotional problems were associated with female avoidance ($P = .083$, respectively, $P = .089$) and dyspareunia ($P = ns$ respectively $P = .094$). In males, role restrictions due to physical problems and health change were related to sexual dissatisfaction ($P = .056$) respectively nonsensuality ($P = .025$).

Conclusions: Overall, Fontan patients have a relatively preserved quality of life and sexual well-being but face more social isolation and bullying during childhood/adolescence than their healthy peers. Sexual problems were mainly associated with physical limitations in males and with psychosocial limitations in females. Finally, sexual dysfunction was more common in older Fontan patients, and future research has to clarify whether progressive attrition of the Fontan circulation affects the patients' QOL and sexual well-being.

KEYWORDS

congenital heart disease, Fontan physiology, quality of life, sexual functioning, special populations

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1 | INTRODUCTION

Since its invention in 1971, the Fontan circulation has become common practice to treat patients with a univentricular heart.^{1,2} With the Fontan operation, the systemic venous return is directed to the pulmonary vascular bed without help of a subpulmonary ventricle. The Fontan circulation is characterized by chronically increased central venous pressure and restricted ventricular filling due to the passive pulmonary blood flow, and an increased ventricular after load, caused by the coupling of the systemic and pulmonary circulation.^{3,4} Various modifications in surgical technique and perioperative care have been developed over the past decades, and short term outcome has significantly improved.⁵ With a growing cohort of patients with a Fontan circulation now reaching adolescence and early adulthood, functional capacity and QOL has become of increasing interest.

Previous studies showed that patients with a Fontan circulation have an impaired exercise tolerance (around 60% of healthy subjects⁶), and are prone to develop various complications, including pulmonary and cardiac disease,⁷ as well as anxiety and depression.^{3,8} The number of health issues in children is previously recognized to be related to the chance of peer rejection and bullying, thereby putting the Fontan patients at risk for social isolation.⁹ However, QOL in Fontan patients seems in general well preserved, with 90%-100% of the patients within normal limits.¹⁰

One important aspect of QOL is sexual well-being. Previous studies showed that adolescents and young adults with congenital heart diseases have increased concerns regarding fertility, inheritability, and pregnancy, may experience a broad range of sexual problems and might lag behind in psychosexual development.^{11,12} Although these studies have included a variety of congenital heart disease, no patients with a Fontan circulation were included. This is unfortunate because the unique physiology of the Fontan circulation justifies special attention for their sexual development and well-being. Important characteristics in this context of QOL/sexual well-being in Fontan patients include restricted cardiac output and exercise tolerance, the chronic systemic venous congestion and potential autonomic dysregulation in response to the decreased cardiac output.¹³ Furthermore, the operations at a young age, frequent hospital visits, and impaired life expectancy might affect patients' QOL and sexual development. Finally, menstrual cycle disorders and fertility problems may influence the patients' sexual well-being.¹⁴

To fill this gap in our knowledge, we decided to perform an explorative pilot study to investigate QOL and sexual function of patients with a Fontan circulation. Its aim was twofold: (1) to get an impression of the QOL and sexual well-being in a sample of the Fontan population, and (2) to generate hypotheses for future research.

2 | METHODS

2.1 | Subjects

In 2012–2013, a cross-sectional study was performed among Fontan patients ≥ 10 years old who were followed at the outpatient clinics of the University Medical Center Groningen, the Netherlands. Of these consecutive patients, all participants ≥ 16 years old were asked to

participate in the current explorative pilot study concerning sexual function and well-being. The institutional ethics committee approved this study. It was conducted in accordance with the declaration of Helsinki and written informed consent was obtained from all study participants.

2.2 | Measures

Patient characteristics were collected from medical records and included, gender, date of birth, cardiac anatomic diagnosis, surgical procedures prior to the Fontan completion, type and date of Fontan completion and current medication use.

To get an impression of QOL and sexual well-being a multimethod approach was used:

1. General health-related QOL (hrQOL) was assessed using the SF-36 questionnaire. The SF-36 questionnaire includes eight health concepts and for each subdomain, scores ranging from 0 to 100 are calculated. The results of the SF-36 questionnaire were compared to a normative sample of 691 females and 372 males, mean age 44.1 years of the Dutch population. Cronbach's alpha of the questionnaire is 0.92 for social functioning, 0.71 for social functioning, 0.90 for role restrictions due to physical limitations, 0.86 role restrictions due to emotional limitations, 0.85 for mental health, 0.82 for vitality, 0.88 for bodily pain, and 0.81 for general health.^{15,16}
2. The Golombok Rust Inventory of Sexual Satisfaction (GRISS) was administered to measure sexual dissatisfaction and problems in heterosexual women and men.¹⁷ Questions regarding contraception, pregnancies, fertility and children were added. After visiting the outpatient clinic, the participating patients were asked to complete the questionnaires at home, allowing for sufficient privacy, and send the completed questionnaires back to the researcher. The results of the GRISS questionnaire of the study population were compared to the reference values of 68 healthy, heterosexual Dutch student couples published by Ter Kuile et al.¹⁸ Mean age of the men in the reference sample was 30.0 ± 8.8 years and 27.5 ± 7.2 years for the women. Cronbach's α of the subsequent subcategories of the GRISS questionnaire, in males and females, respectively, was 0.63 and 0.77 for nonsensuality, 0.84 and 0.85 for dissatisfaction, 0.74 for female dyspareunia, 0.89 for female anorgasmia, 0.78 for male erectile dysfunction, 0.85 for premature ejaculation, 0.71 for avoidance, 0.82 for noncommunication, and 0.94 for infrequency.
3. To generate a clinical impression and hypotheses, ten consecutive patients were asked to participate in semi-structured interviews regarding sexual development and current sexual well-being. These interviews were performed by the first author (D.W.). An outline of the interview structure is presented in Supporting Information file 1.

2.3 | Statistical analyses

Patient characteristics are displayed as mean \pm standard deviation (SD) in continuous variables and as number of patients (percentage of total)

in categorical variables. Results of the questionnaire were compared to the reference values using Levene's test to test the equality of variances and independent *t* test analyses to test the equality of the means. Correlations between the subdomains of the questionnaire on QOL and sexuality were calculated using Pearson's test in normally distributed variables and Spearman's test in skewed variables. A *P* value < .05 was considered significant. All analyses were performed using SPSS for Windows.

3 | RESULTS

3.1 | Subjects

For the questionnaires, 57 patients were asked to participate, of whom 21 patients were eventually included in the current explorative pilot study. The reasons for exclusion were patient refusal (18 patients), no return of the questionnaire (11 patients) or incomplete filled-in questionnaire (7 patients). The participants were 27 ± 5 years old, with a minimum of 19 years and maximum of 44 years. All participants were heterosexual. Of the 14 female respondents, 7 women had an intrauterine device, 1 was on birth control pill, 1 had a birth control implant, 1 woman received birth control shots every 3 months, 1 woman used condom to avoid pregnancy, 1 was sterilized and 2 did not use contraception. Further patient characteristics are listed in Table 1.

For the interviews, ten randomly selected patients were asked. Two patients refused to participate in the interviews because they would rather not talk about sexual well-being. Therefore, the interviews were eventually performed in 8 patients; 3 males and 5 females.

TABLE 1 Patient characteristics (N = 21)

Male, N(%)	7 (33%)
Female, N(%)	14 (67%)
Diagnosis, N(%)	
TA	9 (43%)
DILV	6 (29%)
AVSD/unbalanced VSD	4 (19%)
PA with IVS	2 (10%)
Ventricular morphology, N(%)	
Left dominant	18 (86%)
Right dominant	3 (14%)
Type of Fontan procedure, N(%)	
TCPC lateral tunnel	13 (62%)
Atriopulmonary connection	5 (24%)
TCPC extracardiac conduit	2 (10%)
Bjork modification	1 (5%)
Age at Fontan procedure, years	6.0 ± 4
Current age, years	27 ± 7
Use of beta-blockers, N(%)	7 (33%)
Use of vitamin K antagonists, N(%)	12 (57%)

Abbreviations: (A)VSD, (atrio)ventricular septal defect; DILV, double inlet left ventricle; PA with IVS, pulmonary atresia with intact ventricular septum; TA, tricuspid atresia; TCPC, total cavopulmonary connection.

The short summaries of the interviews are displayed in the Supporting Information file 2.

3.2 | General well-being

The self-reported health related QOL scores of the Fontan patients regarding the domains physical functioning, social functioning, role limitations due to physical problems, role restrictions due to emotional problems, mental health, vitality, pain, and health-change over the last year did not significantly differ from the healthy controls (Table 2). The Fontan patients, however, scored significantly lower on general health than their healthy peers ($t(19) = -3.0$, $P = .008$).

The interviewed patients explained that they currently experience few limitations due to their cardiac condition. They feel that they have a good QOL in general, despite the fact that some (3 out of 8 patients) were not able to work, needed an electronic bicycle (2/8) or not participating in sports due to their physical restrictions (6/8). Conversely, most of the interviewed patients (6/8) had experienced physical limitations during their childhood and/or puberty. This is illustrated by the following statement by patient G:

As a child I realized I was different than the other kids. Now I am used to my limitations, they are a part of who I am and they don't bother me anymore (Female, 25)

and by patient B:

I know that my peers have a higher exercise performance than I have, but nowadays, I do not often get confronted with it. This is in contrast to my childhood, when I was confronted with my restrictions on a daily basis. (Male, 47)

The physical limitations as a child had a clear consequence for almost all interviewed patients; namely the feeling of being an outsider at primary and/or secondary school. Despite the fact that most patients had (at least some) friends, they were from a young age on aware that they were more restricted during exercise and missed more days at school than their peers. Consequently, most of the patients (6/8) experienced forms of bullying during either primary or secondary school.

Patient D:

I liked going to the primary school, but after two months at the secondary school I did not want to go anymore. I was just not part of the group, and therefore an easy victim. I could not participate in sports activities. The rest of the class biked to the city at lunch time, but I always stayed behind because, at that age, nobody will wait for you lagging behind. (Female, 31)

Despite being bullied, none of the patients felt limited to make new friends due to their cardiac condition. More importantly, most of the patients did not report any concerns related to committing to

TABLE 2 Domains of health-related Quality of Life (N = 21)

	SF-36 score		t value (df)	95%CI	P value
	Fontan patients	Z-score			
Physical functioning	73 ± 22	−0.3 ± 0.9	−1.3 (20)	−0.67-0.16	.210
Social functioning	85 ± 16	−0.4 ± 0.8	−1.9 (18)	−0.80-0.04	.076
Role Physical	76 ± 37	−0.2 ± 1.2	−0.9 (20)	−0.76-0.30	.369
Role Emotional	83 ± 31	−0.2 ± 1.1	−1.0 (20)	−0.74-0.26	.331
Mental Health	78 ± 17	−0.1 ± 1.0	−0.4 (20)	−0.55-0.37	.684
Vitality	65 ± 18	−0.2 ± 0.9	−1.1 (20)	−0.68-0.21	.274
Bodily pain	85 ± 18	0.2 ± 0.6	1.8 (20)	−0.04-0.53	.087
General health	59 ± 24	−0.7 ± 1.1	−3.0 (19)	−1.27 to −0.22	.008
Health-change	53 ± 18	0.0 ± 1.1	0.1 (20)	−0.47-0.52	.921

Significant differences ($P < 0.05$) between Fontan patients and reference values are expressed in bold characters.

a friendship or relationship during their adolescence or young adulthood.

3.3 | Sexual function and well-being

Based on the 21 completed GRISS questionnaires, sexual function and well-being in Fontan patients showed no significant differences compared to healthy controls (Table 3, higher scores indicate higher degree of dysfunction). However, in the subdomains concerning female dyspareunia and male erectile dysfunction, 2/21 individual patients had a Z-score > 2 SD compared to their healthy peers, causing significantly larger variances than in the healthy controls ($F = 2.12$, $P = .019$ and $F = 9.30$, $P = .008$). During the interviews, these large interindividual differences were noticed as well. Four patients (2 males and 2 females; 25–30 years old) told that they did not experience any restriction or sexual dysfunction. The other patients (1 male and 3 females, 33–47 years old) reported erectile dysfunction (1/4), a very low self-esteem as

a consequence of childhood experiences (1/4), and severe arrhythmias affecting sexual desire and perseverance (2/4).

Patients B's concerns about erectile dysfunction:

Since my 30th birthday, it is difficult to keep an erection during sexual intercourse. The urologist eventually concluded that it was probably caused by a combination of factors, and one of these was my cardiac condition. Furthermore, the therapeutic options for my problem are very limited due to my cardiac condition. (...) These problems slow me down to take steps in this area with my new partner. (Male, 47)

Regarding the GRISS subdomains infrequency, dissatisfaction, non-sensuality, premature ejaculation (M), and anorgasmia (F), no specific problems were reported during the interviews.

In the subdomain female and male avoidance, the scores of the Fontan patients did not show a significant difference with the normal

TABLE 3 Sexual function and wellbeing

	GRISS score		t value(df)	95%CI	P value
	Fontan patients (N = 21)	Controls (N = 68)			
FEMALES					
Avoidance	5.9 ± 2.8	5.5 ± 1.3	−0.6(109)	−1.52-0.85	.393
Nonsensuality	5.1 ± 1.9	5.5 ± 1.6	0.6(110)	−0.70-1.33	.430
Dissatisfaction	6.6 ± 2.9	6.6 ± 2.2	−0.1(109)	−1.39-1.28	.998
Dyspareunia	6.5 ± 3.3	5.2 ± 1.7	−1.7(109)	−2.60-0.20	.191
Anorgasmia	9.4 ± 3.7	10.0 ± 3.6	0.3(110)	−1.80-2.50	.559
Noncommunication	4.7 ± 2.6	4.5 ± 1.6	−0.2(110)	−1.65-1.38	.746
Infrequency	6.1 ± 2.0	5.3 ± 1.7	−1.9(110)	−2.03-0.03	.111
MALES					
Avoidance	4.3 ± 0.8	4.8 ± 1.0	0.9(102)	−0.42-1.16	.292
Nonsensuality	4.8 ± 1.0	5.1 ± 1.3	0.5(102)	−0.90-1.45	.671
Dissatisfaction	8.2 ± 1.9	8.0 ± 2.7	−0.2(102)	−2.60-2.03	.914
Erectile dysfunction	8.2 ± 3.2	5.5 ± 1.4	−2.0(102)	−3.81--1.32	.096
Premature ejaculation	7.8 ± 2.5	7.7 ± 2.2	−0.04(102)	−1.96-1.88	.867
Noncommunication	3.8 ± 1.3	4.4 ± 1.6	0.7(102)	−0.83-1.82	.441
Infrequency	5.7 ± 1.4	5.1 ± 1.7	−0.8(102)	−2.07-0.90	.457

TABLE 4 Correlations between quality of life and sexual function and wellbeing subdomains

		Physical functioning	Social functioning	Role physical	Role emotional	Mental health	Vitality	Bodily pain	General health	Health change
FEMALES										
Avoidance	R	−0.264	−0.224	−0.413	−0.471	−0.479	0.080	0.132	−0.311	0.329
	P value	.362	.441	.143	.089	.083	.785	.653	.279	.251
Non sensuality	R	−0.113	−0.089	−0.431	−0.399	−0.444	−0.020	0.106	−0.472	0.068
	P value	.701	.763	.124	.158	.112	.945	.717	.089	.817
Dissatisfaction	R	−0.082	0.058	−0.251	−0.217	−0.173	0.116	0.264	−0.314	0.366
	P value	.779	.844	.387	.456	.554	.693	.362	.275	.198
Dyspareunia	R	0.154	0.016	−0.067	−0.465	0.100	0.179	0.163	−0.244	0.159
	P value	.599	.956	.819	.094	.733	.541	.578	.400	.588
Anorgasmia	R	0.077	0.442	−0.150	0.116	0.443	0.374	0.432	−0.380	0.570
	P value	.793	.113	.680	.693	.113	.188	.123	.180	.033
Noncommunication	R	0.190	0.025	0.205	0.025	0.249	0.025	0.286	−0.346	0.162
	P value	.516	.931	.483	.933	.390	.933	.321	.226	.580
Infrequency	R	−0.057	0.154	−0.430	−0.230	−0.325	0.087	0.273	−0.192	−0.135
	P value	.847	.600	.125	.125	.257	.768	.346	.511	.647
MALES										
Avoidance	R	0.539	0.333	0.200	0.200	0.000	0.393	−	0.000	0.566
	P value	.269	.667	.704	.704	1.000	.441		1.000	.242
Nonsensuality	R	0.254	0.544	0.424	0.424	0.188	0.278	−0.566	−0.229	0.867
	P value	.627	.456	.402	.402	.722	.594	.242	.710	.025
Dissatisfaction	R	0.299	0.775	0.799	0.664	0.623	0.207	−0.133	0.802	0.705
	P value	.565	.225	.056 ^a	.150	.186 ^a	.693 ^a	.802	.102 ^a	.118
Erectile dysfunction	R	−0.177	0.775	0.393	0.393	−0.232	−0.371	0.131	0.103	0.494
	P value	.738	.225	.441	.441	.658	.468	.805	.870	.320
Premature Ejaculation	R	−0.104	0.258	−0.033	−0.133	−0.263	−0.499	−0.399	0.020	0.016
	P value	.844	.742	.951 ^a	.802	.614 ^a	.314 ^a	.434	.975 ^a	.977
Noncommunication	R	−0.429	0.577	0.283	0.283	−0.235	−0.463	0.283	−0.135	0.217
	P value	.396	.423	.587	.587	.654	.355	.587	.828	.680
Infrequency	R	0.061	0.577	0.270	0.270	−0.403	−0.324	−0.270	0.081	0.683
	P value	.909	.423	.605	.605	.428	.531	.605	.897	.135

Significant differences ($P < 0.05$) between Fontan patients and reference values are expressed in bold characters.

^aPearson's correlation test.

All other correlations = Spearman's correlations test.

values. However, the two interviewed patients who reported that severe arrhythmias affected their sexual life, indicated that they tend to avoid sexual intercourse. These patients avoided sexual intercourse because they are afraid to instigate or worsen the arrhythmias.

Patient E:

Nowadays I tend to avoid sexual intercourse. Every time we have sex, my arrhythmias start again. It really decreases your desire to have sex. I can not enjoy it anymore, because I can not relax, constantly thinking about my cardiac condition. (Female, 46)

Concerning the GRISS subdomain noncommunication, no problems were reported during the interviews about the current relationships of the patients. However, three out of eight interviewed patients reported that they found it difficult to tell about their cardiac condition to a new partner, mostly because they were afraid of their reaction. They thought their new partner would be scared to share a life with someone with a severe cardiac condition, restricted life expectancy, frequent hospital visits and restricted exercise limitations along with sexual impairment (in 1 male patient).

Significant correlations between the QOL subdomains and GRISS subdomains are demonstrated in Table 4.

3.4 | Fertility, pregnancy, and children

Three out of the fourteen females who completed the questionnaires had been pregnant. Two of these women sought for help at the fertility clinic. Together, the women had four pregnancies; one ended in a miscarriage (< 20 weeks pregnancy) and the other three babies were born prematurely (at 27, 31, and 35 weeks pregnancy). Of the latter three babies, one died after two months due to meningitis and the other two are still alive. Eight of the female participants reported that they wish to have children in the future, of whom five females were discouraged by their physician to have children due to their heart condition.¹⁹

Of the five interviewed women, two reported a low self-esteem due to the fact that they could not, or are discouraged to, become pregnant.

Patient Fs' answer to the question if she has to offer enough as a partner:

I find that very difficult. My partner and I cannot have any children, because of my cardiac condition. I struggle with the fact that I cannot offer him children, while another woman could. It would have been easier for me if he also had a fertility problem. (Female, 33)

Based on the questionnaires, two of the males reported that they established a pregnancy in a woman, both without any help of fertility improving therapy. In both cases, the child was born healthy, without any heart defect. Three of the other males reported a desire for children in the future. None of males were, because of their heart condition, discouraged by their physician to get children.

During the interviews, none of the males was concerned about fertility or raising children.

4 | DISCUSSION

In this study, we found Fontan patients to report a health-related QOL and sexual well-being generally comparable to normal values. However, during childhood, most patients experienced physical limitations and the feeling of being an outsider. Regarding sexual well-being, large interindividual differences were noted especially in the subdomains dyspareunia and erectile dysfunction of the questionnaire, and regarding erectile dysfunction, avoidance, and self-esteem during the interviews. Based on this study, some impressions could be sketched:

Fontan patients face daily restrictions and co-morbidities, associated with the Fontan circulation. Despite these restrictions, this study confirmed that these patients seem to experience a generally well preserved health-related QOL. However, the interviewed patients pointed out those physical restrictions were more prominent during childhood, where they consequently felt an outsider in their peer-group and were often bullied. The number of Fontan patients who had the experience of being bullied seems rather high compared to a recent study on school-related adjustment, which demonstrated that 9% of the adolescents with congenital heart disease had the experience of being bullied.²⁰ However, the cardiac malformation of the majority of the adolescents in the latter study was completely surgically corrected. Whereas the study by Casey et al showed that children in whom the cardiac disease was not completely corrected, were more withdrawn and had more social problems.²¹ Potentially, overprotection from parents and the degree in which the disease interfered with participation in school or play affected their sense of normalcy.^{22,23} Furthermore, Fontan patients often face multiple health issues, putting them at risk for peer rejection and bullying.⁹ To our knowledge, the high prevalence of social isolation and bullying of Fontan patients is not previously recognized. These are important issues needing attention because they can influence adult self-esteem and mental health outcomes.²⁴ In the near future, more prospective pediatric studies to investigate the hypothesis based on this pilot study whether Fontan patients indeed experience significantly more social isolation and bullying are needed. Moreover, the benefit of promotion of self-efficacy and specific skills training, might be of great interest to help the patients with these issues.^{25,26}

The operation(s) at a young age and the abnormal circulation can potentially affect both psychological and physical aspects of the patients' well-being. Psychologically, the impaired life expectancy, body- and self-esteem, performance anxiety, and impaired fertility might be important factors. Physically, the increased central venous pressure, limited exercise capacity and medication use might impair sexual function. In general, sexual function and well-being seemed well preserved in this

study. Some of the patients experienced a limited exercise capacity during sexual intercourse, but this did not impact their sexual well-being. This is in line with results from previous studies on a variety of diseases, including congenital heart disease.²⁷ Conversely, large interindividual differences were noted and sexual dysfunction was described by several patients on an individual level. Sexual impairment reported in this study included erectile dysfunction, low body- and sexual self-esteem and avoidance of sexual intercourse. In previous studies investigating patients with other congenital heart diseases, it has been reported that 10% of the males suffer from erectile dysfunction, potentially caused by impaired cardiac output and restricted functional capacity.^{28,29} Furthermore, a recent study demonstrated that medication use, including spironolactone and digoxin, significantly affected sexual function in men with congenital heart disease.³⁰ In addition, in Fontan patients, erectile dysfunction might be provoked by a dysregulation of the autonomic nervous system, chronic systemic venous congestion and endothelial dysfunction.^{31,32} One of the three males who were interviewed was unable to maintain an erection during sexual intercourse. This problem affected his sexual life and he was very limited in the pharmacological therapeutic options (eg, Sildenafil) for his problem.

In women with congenital heart disease, sexual dysfunction, including dyspnea, arrhythmias, fatigue and syncope, and lower self-esteem are previously recognized.^{27,33} In this study, the females reported predominantly psychological components which affected their esteem of their sex life and partnership (patients C and F). Their main concern being their inability to offer children to their partners. High concerns regarding fertility and pregnancy are also reported in patients with other congenital heart defects.³⁴ However, those patients were more concerned about their own health and their ability to carry the baby to term, whereas women with a Fontan circulation were discouraged to become pregnant and were concerned that their partner might consider having children with another woman.

In addition, we examined the correlation between the health-related QOL and sexual functioning. Correlations between health-related QOL and male-specific subdomains of the GRISS questionnaire as well as the subdomain female anorgasmia were primarily driven by two outliers (one male, one female). Conversely, despite the small sample sizes we were able to demonstrate that both mental health and role restrictions due to emotional problems show a trend toward significance (P value $< .10$) with female avoidance and dyspareunia. This corresponds with a recent report by Ghizzani et al, showing that sexual pain in women can affect the women's sense of well-being.³⁵

Interestingly, four interviewed patients reported no sexual dysfunction, suggesting that sexual function can be well preserved in the Fontan circulation. In the Fontan circulation, progressive deterioration of the circulation has been suggested, also known as Fontan attrition, manifesting itself by various circulatory complications, including gastroenteric complications, arrhythmias, and deteriorating functional capacity.³ The four patients who did not report sexual dysfunction, were the youngest interviewees and in none of these patients adverse sequelae of the Fontan circulation had developed (yet). Therefore, gradual attrition of the Fontan circulation may be associated with progressive sexual (dys)function of the Fontan patients over time. This is

also confirmed by patients C and E, who explained that their arrhythmias and the anxiety for a new onset of arrhythmias severely impact their sexual desire and activity, as well as by patient B, who suffers from erectile dysfunction from the age of thirty.

In the most recent guidelines provided by the American College of Cardiology (ACC) it is stated that sexual activity, contraception and pregnancy issues are key issues to monitor in patients with tricuspid atresia/Fontan circulation.³⁶ However, health care providers might be hesitant to start a conversation on these topics, due to lack of time in clinical practice, limited knowledge or fear for upsetting the patient.^{37,38} The ACC/American Heart Association have introduced a scientific statement with general recommendations to help the clinician with this clinical important issue.³⁹

The results of this study need to be interpreted in the context of several limitations. First, it is important to notice that the patients were very well-spoken regarding their experiences as a child or youngster with a congenital heart defect. However, when the interviewer addressed their current status of "what they have made of their lives" or "how they address their problems," the patients found it more difficult to find words and describe their feelings and functioning. Second, this study included a sample of 21 patients who returned the questionnaires, out of 62 patients' ≥ 16 years old in our Fontan cohort. This causes a potential selection bias and might have caused an underrepresentation of psychosocial or sexual problems. To highlight the relevance of sexuality as an important theme, we focused in our design on the differences with the general population. However, we want to emphasize that the Fontan population not only resembles the general population in many ways, but that within our patient population an attribution error may have occurred. When there is one clear difference between the patient population and the general population (for instance cardiac anatomy), all kind of other differences might be attributed to this feature, whether or not appropriate. Because we used an exploratory design, we could not correct or compensate for this well-known psychological phenomenon. Third, the Cronbach's alpha for the subscales nonsensuality in males is rather low and the results regarding this subscale have to be interpreted with care. Finally, the comparison between Fontan patients and the normative sample size of healthy couples who filled in the GRISS questionnaire, as well as the correlations analyses have to be interpreted with care due to the relative small sample sizes and lack of matched comparison data. However, this study was not designed to collect quantitative data, but aimed at generating first impressions and hypotheses regarding QOL and sexual well-being in Fontan patients. Future studies have to be conducted to investigate the prevalence of sexual problems and to identify whether sexual well-being of Fontan patients differs from patients with other congenital heart defects.

In summary, as a result of this explorative pilot study, we found several general themes regarding single ventricle patients and social/sexual development and well-being. In general, the patients with a Fontan circulation seem to have a relatively preserved quality of life but face more social isolation and bullying during childhood/adolescence than their healthy peers. Regarding sexual dysfunction, Fontan patients seem to adapt fairly well to their physical restrictions and only a minority of the patients experienced problems. Possibly, there is a male/

female difference regarding sexual well-being in patients with a Fontan circulation. In males, physical limitations predominate and in females psychosocial limitations predominate. Finally, sexual dysfunction seems more common in older Fontan patients, and future research has to clarify whether progressive attrition of the Fontan circulation affects the patients' QOL and sexual well-being.

CONFLICT OF INTEREST

The University Medical Center Groningen has received fees for consultancy activities of RMF Berger for Actelion, Pfizer, Bayer Lilly, and GSK outside the content of this manuscript.

AUTHORS CONTRIBUTIONS

Study design: Wolff, van de Wiel, van Melle, Pieper, Berger, Ebels, Schultz

Data acquisition: Wolff, Keizer

Data analysis/interpretation: Wolff, Wiel, Melle, Berger, Ebels, Schultz

Drafting article: Wolff, Keizer

Statistics: Wolff

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SUPPORTING INFORMATION

Additional Supporting Information may be found online in the supporting information tab for this article.

File S1. Interview structure.

File S2. Interview summaries.

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